The morphologic nature of noncommitted ventricular septal defects in specimens with double-outlet right ventricle

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Background: Lev's contribution to the understanding of the morphology of hearts with double-outlet right ventricle and the surgical feasibility for correction is important and remains in current use. However, the term *noncommitted ventricular septal defect* remains enigmatic. The aim of this study was to elucidate the morphologic nature of the noncommitted ventricular septal defect in view of its surgical implications.

Methods: We examined 67 specimens with double-outlet right ventricle, focusing on the relationship of the ventricular septal defect to the semilunar orifices.

Results: The defect was subaortic, subpulmonary, or doubly committed in 55 specimens. In a further 8 specimens, the defect opened into the outlet portion of the right ventricle, but the distance between the ventricular septal defect and the semilunar orifice was extensive, either because of extreme dextroposition of the aorta or a broad ventriculoinfundibular fold, which, in some cases, was associated with a long-outlet septum. A truly noncommitted ventricular septal defect was found in the inlet in the remaining 4 specimens. An atrioventricular septal defect without extension to the outlet was present in 3 cases, and a ventricular septal defect limited to the inlet was found in another case. The ventriculoinfundibular fold, part of the outlet septum and septal band or septomarginal trabeculation, had fused to form a crestlike structure. The septomarginal trabeculation is a useful landmark in the right ventricle to differentiate the inlet from the outlet in different forms of double-outlet right ventricle.

Conclusion: We do not suggest to discard the Lev terminology but rather to differentiate the noncommitted ventricular septal defect into 2 types: the truly noncommitted defect of the inlet type and the not-directly-committed defect, which does open into the outlet portion of the right ventricle. The implication for the surgeon is 2-fold. The tricuspid valve or right part of the atrioventricular valve is interposed between the noncommitted ventricular septal defect and the semilunar orifice. The not-directly-committed defect opens into the outlet portion of the right ventricle but is not directly subaortic or subpulmonary.

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Double-outlet right ventricle (DORV) is an abnormal ventriculoarterial connection of the heart in which both great vessels arise completely or predominantly from the right ventricle (RV), and it comprises several different cardiac malformations, of which a ventricular septal defect (VSD) is an integral feature.[1-14] Although the abnormal location of the aortic orifice in the outlet portion of the RV is an obligatory feature of DORV, the accompanying VSD is not defined as being in the outlet. Morphogenetically, a DORV ventriculoarterial connection is not an entity, although all specimens have in common a deviation of the outlet septum with subsequent disruption of the supraventricular crest.[15] In the normal heart the supraventricular crest is a muscular structure that separates the outlet of the RV from the tricuspid valve. Disruption of this structure usually causes a VSD in the outflow portion of the septum. There is, however, contention with regard to the VSD being in the outflow portion of the septum in some cases of DORV. These are the so-called noncommitted VSDs, a term that was introduced by Lev and colleagues.[3] Noncommitment meant that the VSD was related to neither semilunar valve, being separated from both by considerable muscle. In addition, their concept of noncommitment was meant to have surgical importance without precisely defining the anatomic substrate. Furthermore, the concept of noncommitment as was then proposed has little bearing on the feasibility of univentricular or biventricular repair in this era. Anderson and colleagues,[13] in describing the surgical anatomy of the VSD in the DORV state, found that the noncommitted variant includes a defect embedded within the musculature of the inlet or apical ventricular septum or as part of an atrioventricular septal defect (AVSD) but also in the outlet, in which case the direct relationship of the VSD to the semilunar valve is disturbed because of the length of the subarterial infundibulum. Stellin and coworkers[16] extended the meaning of noncommitment to include structures obstructing the space between the VSD and orifice of the great vessel, such as the leaflets of a straddling atrioventricular valve. Belli and associates[17] recently stated that a noncommitted VSD is not an anatomic definition.

The aim of the study was to elucidate the morphologic nature of the noncommitted VSD in view of its surgical implications.

Materials and Methods

Of the 80 available specimens, 13 were not suitable for examination because of inappropriate sectioning, damage to structures caused by frequent examinations, or alteration of intracardiac structures by previous operations. In this study we included specimens in which both semilunar orifices are connected to the RV totally or exhibited more than 50% overriding of the ventricular septum. The presence of fibrous continuity between a semilunar orifice and an atrioventricular orifice did not exclude entry into the series. An analysis was performed for the detection of other abnormalities of the heart. One of the borders had to be fibrous, abutting on the central fibrous body, to determine whether the VSD was perimembranous.[18] The walls of both outflow tracts were then examined, as was proposed by De la Cruz and colleagues[11] and elaborated on by us.[19] By using the septal band or septomarginal trabeculation (SMT) as a marker, outflow tracts can be classified as having anteroposterior or side-by-side relationships in most cases. The relationship is indeterminate if the SMT is incomplete. In case of the VSD opening into the outlet portion of the RV, the outflow tract into which the VSD opened was noted. In this way the VSD was categorized as being subaortic or subpulmonary. If the outlet septum was rudimentary, with fibrous continuity between both semilunar valves, the VSD was doubly committed. Features contributing to an outlet VSD not being in close proximity to one or both semilunar orifices or structures obstructing the space between were noted. Specimens with these features were defined as having a not-directly-committed VSD.[20]

A VSD that did not extend into the outlet portion of the RV or that was isolated from the outlet was defined as being noncommitted.[14,20,21]

TABLE 1. Morphological characteristics of the VSD

Total No. of specimens	67
Position	
Subaortic	25
Subpulmonary	24
Doubly committed	6
Not directly committed	8
Opening into subaortic outflow tract	7
Opening into subpulmonary outflow tract	1
Non committed	4
Inlet type	1
AVSD type without extension to outlet	3

TABLE 2. Most frequently occurring forms of associated congenital heart disease*

Total No. of specimens	67
Specimens with associated congenital heart disease	44
Left ventricular hypoplasia	25
AVSD	6
Valvular pulmonary atresia	2
Atrioventricular discordance	2
Straddling mitral valve	2
Straddling tricuspid valve	2
Accessory mitral valve tissue	2
Mitral valve malattachment	2

*Many specimens had more than one form of associated congenital heart disease.

Results

Of the remaining 67 specimens, there were 55 in which the VSD was subaortic, subpulmonary, or doubly committed

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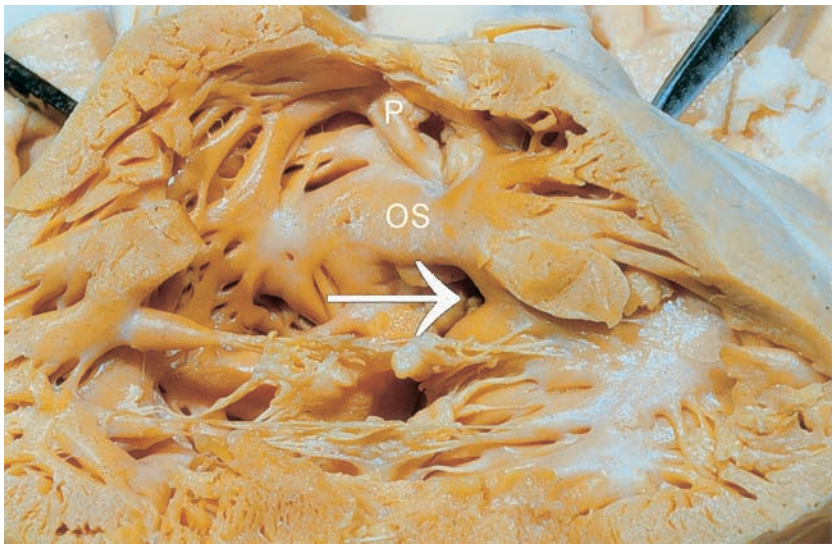

Figure 1. Subaortic VSD. Right ventricle in specimen with DORV with an anteroposterior outflow tract. The Y of the septomarginal trabeculation (*arrow*) cradles the VSD. The *arrow* overlies the aortic orifice. The subpulmonary outflow tract (*P*) lies anterior to the outlet septum (*OS*) and is not stenotic.

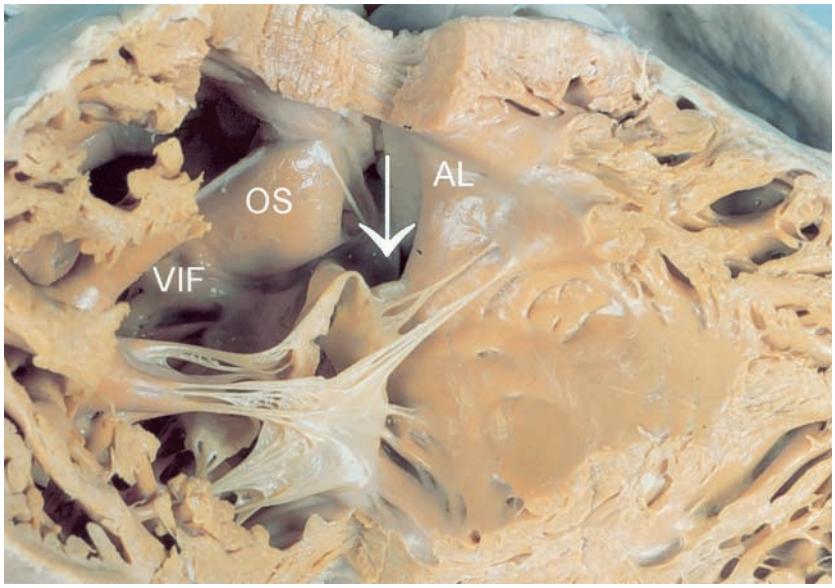

Figure 2. Subpulmonary VSD. Right ventricle in specimen with DORV with side-by-side outflow tracts. The Y of the SMT (*arrow*) cradles the VSD. The anterior limb (*AL*) of the SMT is not closely related to the outlet septum. The VIF is connected to the outlet septum (*OS*). The lateral outflow tract, which lies to the left and somewhat anterior on the figure, is subaortic.

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(Table 1). In Table 2 association with congenital heart disease is listed. An example of a subaortic VSD is demonstrated in Figure 1, and a subpulmonary VSD is demonstrated in Figure 2. Except for specimens with an AVSD, these 55 specimens were not subjected to further study.

In one specimen (Figure 3) there was one VSD limited to the inlet with a complete muscular rim, which was therefore classified as noncommitted. The ventriculoinfundibular fold, part of the outlet septum, and the SMT were fused to form a structure similar to a supraventricular crest. The aorta was to the right, side by side with the pulmonary artery with double conus, and was completely within the RV. Mitral atresia and severe left ventricular hypoplasia were added non–right ventricular features that contributed to the

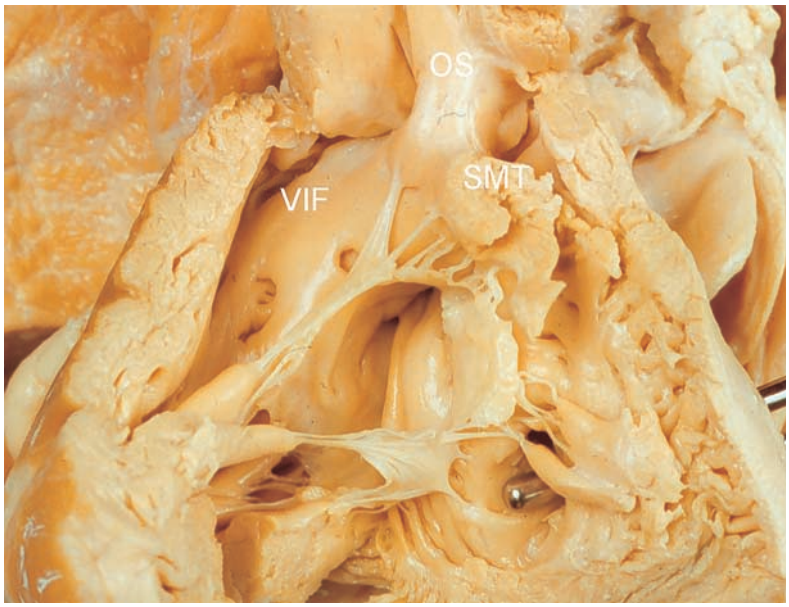

Figure 3. Noncommitted VSD. Right ventricle in specimen with DORV. The probe lies through the noncommitted muscular inlet VSD, which lies below the tricuspid valve on this figure and has no relationship with the septomarginal trabeculation (*SMT*), which has been sectioned. *VIF*, Ventriculoinfundibular fold; *OS*, outlet septum.

TABLE 3. Features of DORV specimens with not-directly-committed VSDs

Specimen No.	7	9	10	13	14	54	64
Aortic orifice to the right, side by side		+				+	
Aortic orifice to the right, posterior	+		+	+	+		+
VSD, perimembranous	+	+	+	+	+	+	+
Percentage override of orifice over septum	100	100	80	100	80	100	100
Presence of double conus	+	+		+		+	+
Subpulmonary stenosis; pulmonary valvular stenosis/atresia	+	+			+		
Subaortic stenosis							+
Associated congenital heart disease	+	+	+			+	+
Hypoplasia, left ventricle		+	+				+
Accessory mitral valve tissue	+						
Extensive VIF	+	+	+	+	+	+	
Long-outlet septum	+	+	+				
Extreme dextroposition of aorta	+	+		+			+
Straddling mitral valve						+	
Straddling tricuspid valve						+	
RV features preventing biventricular repair							+
Non-RV features preventing biventricular repair		+					+
Feasibility of biventricular repair	+		+	+	+	+	

RV, Right ventricular; *VIF*, ventriculoinfundibular fold.

nonfeasibility of biventricular repair in this specimen. There were 8 specimens in which the VSD opened into the outlet of the RV but were not directly committed, the features of 7 of which are listed in Table 3. In Figure 4 a broad ventriculoinfundibular fold is demonstrated. In Table 4 a detailed list of features of all 6 specimens with an AVSD is given.[22] Typically, the VSD associated with an AVSD is in the inlet, whereas in cases of DORV the VSD is in the outlet portion. In these hearts with an AVSD and DORV, we found a spectrum of combinations, all with inlet VSD and varying degrees of extension to the outlet. There were 3 specimens with minimal extension to the outlet. Of the other 3 specimens with AVSD, the VSD did extend into the outlet, with 2 being subaortic VSD and one being not

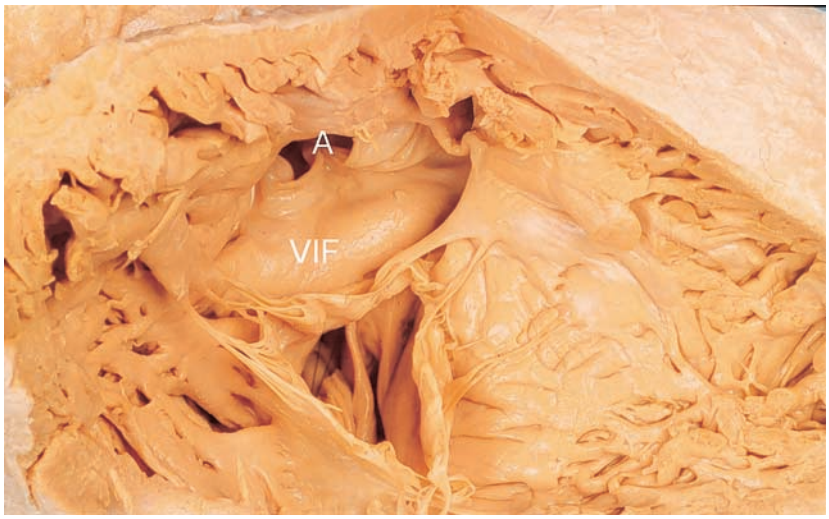

Figure 4. Not-directly-committed VSD. Right ventricle in specimen with DORV. Aortic orifice (*A*) with broad ventriculoinfundibular fold (*VIF*) beneath it. On the figure the VSD, which opens into the outlet portion, lies to the right of the aorta and VIF and is cradled by the septomarginal trabeculation.

TABLE 4. Features of DORV specimens with AVSD

Specimen No.	2	22	26	58	62	63
Aortic orifice to the left, anterior						+
Aortic orifice to the left, posterior/side by side				+		
Aortic orifice to the right, side by side					+	
Aortic orifice to the right, posterior	+	+	+			
Percentage override of orifice over septum	100	100	80	100	100	100
Presence of double conus		+		+		+
Subpulmonary stenosis; pulmonary valvular stenosis/atresia	+		+	+	+	+
Associated congenital heart disease	+		+	+		+
Hypoplasia, left ventricle	+		+			+
Atrioventricular discordance				+		+
AVSD inlet VSD with extension to outlet	+		+	+		
Outlet VSD, subaortic	+		+			
Outlet VSD, not directly committed				+		
AVSD inlet VSD without extension to outlet		+			+	+
Extensive VIF				+	+	+
Obstructive septomarginal trabeculation						+
Extreme dextroposition of aorta	+			+		
RV features preventing biventricular repair		+			+	+
Non-RV features preventing biventricular repair	+		+			
Feasibility of biventricular repair				+		

RV, Right ventricular; *VIF*, ventriculoinfundibular fold.

directly committed (Figure 5). In 22 specimens aberrant chordae tendineae were present in the outlet but did not obstruct the space between the VSD and the semilunar valve. Although this is a morphologic study, we also examined the specimens with a view to the feasibility of biventricular repair, and in Tables 3 and 4 we have noted RV and non-RV features that might prevent this.

Discussion

Lev and colleagues's[3] contribution to the understanding of the morphology of hearts with DORV and the surgical feasibility for correction is important and remains in current use. However, we agree with Belli and coworkers[17] that the term *noncommitted VSD* is not an anatomic definition because the description by Lev and colleagues[3] of separation

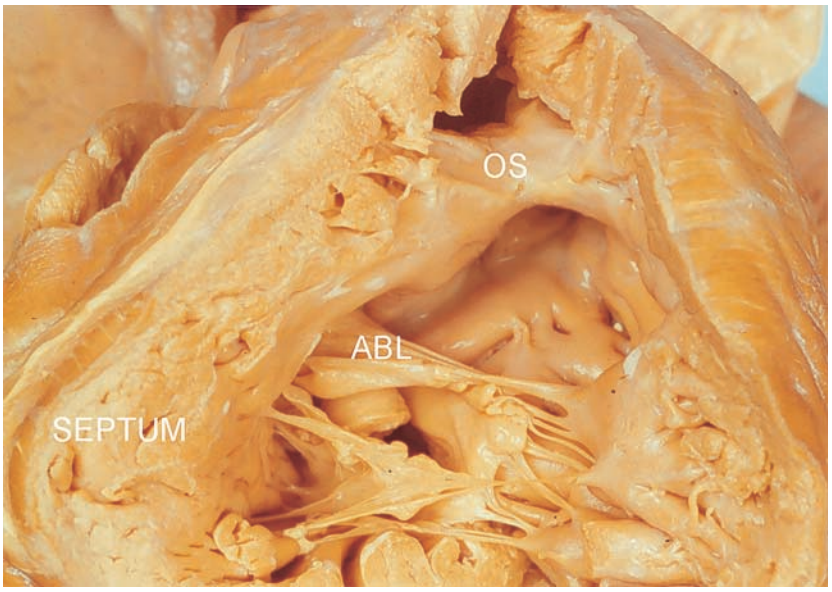

Figure 5. Not-directly-committed VSD. Left-sided right ventricle with DORV in specimen with atrioventricular discordance, complete atrioventricular septal defect in which the anterior bridging leaflet (*ABL*) is seen passing through the VSD. The VSD has an inlet and outlet component. There is some distance from the outlet component of the VSD to the aortic orifice, which lies below the outlet septum (*OS*) on this figure.

of the semilunar valve from the VSD by considerable muscle is neither exclusive nor comprehensive of a number of conditions.

In this study we found that in 12 of 67 specimens, the VSD did not have a direct relationship to one or both semilunar valves. Of these 12 specimens, there were 8 in which the VSD opened into the outlet portion and 4 into the inlet portion of the RV. In addition, aberrant chordae tendineae were found in 22 cases, but these were not found to obstruct the space between the VSD and the great vessel orifice.

For morphologic purposes, the terms *inlet* and *outlet* define the position of the VSD, even in the setting of DORV. The septal band, or SMT, is a useful landmark in the RV to differentiate the inlet from the outlet, as was described by Lev and colleagues[3] in different forms of DORV. The inlet VSD can be further differentiated into those that are and are not associated with an AVSD.[14,21]

For surgical purposes, it is useful to denote the inlet VSD as a separate type of noncommitted VSD when adhering to the Lev terminology because the connection between the VSD and the semilunar valve will have to bypass the tricuspid valve or the right atrioventricular valve. Defining the VSD in terms of commitment has surgical implications rather than a strict anatomic basis and can be superimposed on the morphologic definition. We therefore do not suggest disbanding the Lev terminology but suggest a differentiation of the noncommitted VSD into 2 types, the truly noncommitted VSD of the inlet type and the not-directly-committed VSD,[20] which does open into the outlet portion of the RV, also to accommodate current surgical practice.[17,21,23-27] In fact, Barbero-Marcial and associates,[21] in their article on biventricular repair of DORV, have reserved the term *noncommitted* for an inlet VSD below the septal band or its divisions. To bypass the tricuspid valve or the right atrioventricular valve, they describe a technique with 2 patches and in some cases reduce the tricuspid valve with reinsertion of papillary muscles at the patch. In case of an inlet VSD of the atrioventricular type, in which there is no extension to the outlet, the creation of a hole in the outlet portion of the septum adjacent to the perimembranous area, corresponding to the interventricular foramen in embryologic development and closing the inlet part of the VSD, is another theoretic possibility.

The key feature of a noncommitted VSD is that it is an inlet VSD. The key feature of a not-directly-committed VSD is that it is in the outlet portion of the RV but that the presence of an extensive ventriculoinfundibular fold, long-outlet septum, extreme dextroposition of the aorta, or aberrant chordae tendineae in the outlet of the RV disturb the direct relationship of the VSD to the semilunar orifice.

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